***Case Report***

# Aggressive Recurrent Urachal Adenocarcinoma : A Case Report And Review Of The Literature

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**Abstract**

Recurrent urachal adenocarcinomas are rare. It is commonly associated with poor prognosis. A 51-year-old woman underwent a partial cystectomy for urachal cancer in 2014. She was well throughout follow-up with annual cystoscopies. She presented with a 1-month history of rapidly progressive suprapubic mass and hematuria. A contrasted computed tomography scan of the thorax, abdomen and pelvis showed a large mixed solid cystic tumour at the right lumbar region, suprapubic area and also at the pouch of Douglas. She refused surgical extirpation and now on chemotherapy. Urachal adenocarcinoma is rare, and resection is commonly advocated; usually, no standard adjuvant therapy is advocated. Recurrent cancers represent a management dilemma, and no standard follow-up protocols exist.

**Keywords:** Urachal; adenocarcinoma; recurrent; partial cystectomy

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## Introduction

Urachal adenocarcinoma is rare. Hue and Jacquin first described it in 1863[1]. It accounts for 0.5% of all primary bladder cancers, and most commonly occurs in males[1]. It arises from the urachus, a vestigial remnant of the allantois that extends from the dome of the bladder and to the umbilicus. Urachal cancer has a dismal prognosis. We present a woman with urachal cancer that recurred four years post-partial cystectomy. The patient was not treated with any adjuvant chemotherapy. She presented with a large abdominal mass and multiple visceral metastases. Our case illustrates the aggressive nature of this recurrence despite long term remission, the usage of folinic acid, 5-fluorouracil and oxaliplatin-based (FOLFOX) chemotherapy for this situation and review of the existing literature.

## Case report

A 51-year-old woman presented with a suprapubic mass, lower abdominal discomfort and hematuria for three months. Further history did not reveal any other significant details. She underwent a cystoscopy that showed a large mass at the bladder's dome with a thin remnant traced down to the bladder. She underwent a biopsy that showed urachal adenocarcinoma and proceeded with partial cystectomy. She did not experience any immediate complications post-operatively. No adjuvant therapy was administered for her.

She was well throughout four years of follow-up with serial cystoscopies performed for her. After which she complained of hematuria and a suprapubic mass for a month duration. A contrasted computed tomography scan of the thorax, abdomen, and pelvis

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was performed to investigate the suprapubic mass further. It showed several nodules seen in both lungs, largest at the right apex measuring 8 x 12 mm with small pleural effusion, large mixed solid cystic tumour at the right lumbar region measuring 13.6 x 12.9 x 15.0cm (Figure 1) with large lobulated solid with cystic mass lesion at the suprapubic region measuring 8.5 x 8.3 x 12.5 cm (Figure 2) and also large loculated solid cystic mass at pouch of Douglas measuring 6.3 x 10 x 2.7 cm. Ultrasound-guided needle biopsy of the suprapubic mass revealed recurrent urachal adenocarcinoma with similar tumour morphology as the previous resected cancer.

After a multidisciplinary meeting to discuss further management, she was started on chemotherapy. The regime of choice was FOLFOX. A repeat resection was offered to the patient however, she declined. She has completed 3 out of 6 cycles of chemotherapy thus far.

**Table 1: The MD Anderson Cancer criteria for the diagnosis of urachal cancer**

| | |
|---|---|
| Main criteria | |
| | Location in the bladder dome or elsewhere in the midline of the bladder |
| | Sharp demarcation between tumour and normal surface epithelium |
| Supportive criteria | |
| | Enteric type histology |
| | Absence of urothelial dysplasia |
| | Absence of cystitis cystica or cystitis glandularis transitioning to tumour |
| | Absence of primary adenocarcinoma of another origin |

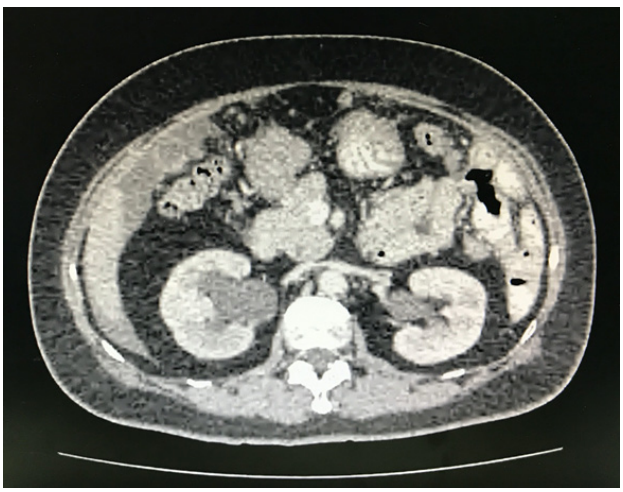

**Figure 1**: Mixed solid cystic tumour at the right lumbar region

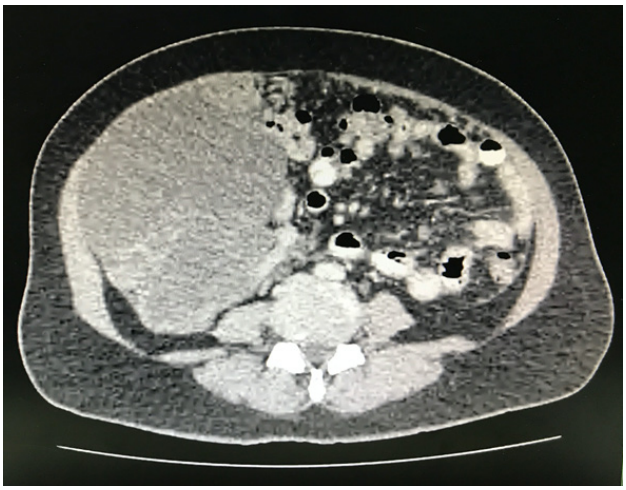

**Figure 2**:Large mixed solid cystic tumour at the right and suprapubic region

## Discussion

Anatomically, the urachus is a vestigial muscular fibrous band of tissue located in the space of Retzius. It is surrounded anteriorly by the transversalis fascia and posteriorly by peritoneum.[2] If remnants of the allantois remain within the ligament, they may develop into cysts and epithelial neoplasms. Urachal remnants have been identified in one-third of cases in post mortem studies; in the dome and anterior wall commonly and rarely in the posterior wall of the bladder.[3]

Patients usually present late, and symptoms are generally non-specific because the disease arises from outside of the bladder. Symptoms typically occur after invasion into the bladder and can manifest as hematuria and irritative voiding.[4] Standard hematological investigations are applied, including cystoscopy with or without a biopsy. Imaging modalities are utilised to rule out metastases. The utility of tumour markers may be of as with any other enteric malignancies. Elevations in carcinoembryonic antigen (CEA), Ca-125 and Ca 19-9 have been reported in some cases and changes in the value used to predict response to primary treatment.[5]

Numerous criteria exist to diagnose urachal cancers, whereby the widely used is based on the MD Anderson Cancer Center (MDACC) (Table 1). It includes location, the tumour, demarcation of epithelium and enteric histology[6]. The mainstay of treatment for these tumours is usually open cystectomy or partial cystectomy with en-bloc resection of the median umbilical ligament with clear margins. As for adjuvant therapy, no standard regimen is available, and usage of chemotherapy is generally based on case reports and single-institution experience.

Recurrent urachal cancers occur up to 20% of cases, and similar to primary cancer treatment, surgical extirpation is advocated with it being the most crucial factor for prolonged survival.[7] Regarding chemotherapy usage, as with primary urachal cancers, the regimen to be utilised is based mainly on case reports. Yasuyoshi reported the usage of combination chemotherapy of gemcitabine and cisplatin in a gentleman who developed recurrence five months after primary surgery. The patient refused the proposed full course of treatment due to side effects and death ensued 16 months after diagnosis.[8]

Usage of the FOLFOX regime has been described in metastatic primary urachal adenocarcinoma; however, not many report its usage in recurrent disease. It is postulated that due to its enteric cellular similarities, FOLFOX might be useful in prolonging survival for our patient, and we will continue to update its outcome.

## Conclusion

Recurrent urachal adenocarcinomas are rare and aggressive. The mainstay of treatment is surgical extirpation, and in the absence of standard guidelines, chemotherapy regimens are purely experimental thus far. Usage of FOLFOX based chemotherapy might prove to be effective.

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**Authors' contribution**: ZAZA owned the idea of writing this manuscript and prepared the table and figures. MAHS wrote the manuscript. FH became the corresponding author and replied to the editor. ZZ provided expert opinion and revised the final manuscript. All authors approved the final manuscript.

## References

1. Ghazizadeh M, Yamamoto S, Kurokawa K. Clinical features of urachal carcinoma in Japan: review of 157 patients. *Urol Res*. 1983;**11**(5):235-8. https://doi.org/10.1007/BF00272286
2. Scabini S, Rimini E, Romairone E, et al. Urachal tumour: case report of a poorly understood carcinoma. *World J Surg Oncol*. 2009;**7**:82. https://doi.org/10.1186/1477-7819-7-82
3. Gopalan A, Sharp DS, Fine SW, et al. Urachal carcinoma: a clinicopathologic analysis of 24 cases with outcome correlation. *Am J Surg Pathol.* 2009;**33(**5):659-68. https://doi.org/10.1097/PAS.0b013e31819aa4ae
4. Hadian B, Khodadadi B, Rezaei N, Ghanadi K. A very rare case report of sigmoid adenocarcinoma with involvement of the bladder at the time of initial diagnosis. *Bangladesh Journal of Medical Science.* 2019;**18(**4):827-829 https://doi.org/10.3329/bjms.v18i4.42914
5. Guarnaccia S, Pais V, Grous J, Spirito N. Adenocarcinoma of the urachus associated with elevated levels of CA 125. *J Urol.* 1991;**145**(1):140-1 https://doi.org/10.1016/S0022-5347(17)38271-X
6. Siefker-Radtke A. Urachal carcinoma: surgical and chemotherapeutic options. *Expert Rev Anticancer Ther.* 2006;**6(**12):1715-21. https://doi.org/10.1586/14737140.6.12.1715
7. Kawakami S, Kageyama Y, Yonese J, et al. Successful treatment of metastatic adenocarcinoma of the urachus: report of 2 cases with more than 10-year survival. *Urology.* 2001;**58(**3):462. https://doi.org/10.1016/S0090-4295(01)01259-6
8. Miyata Y, Sagara Y, Matsuo T, et al. Response of recurrent urachal cancer to gemcitabine and cisplaatin therapy: A case report and literature review. *Anticancer Res*. 2011;**31**(31):2335-8.